Summary

A further case of acute phlegmonous caecitis is described. The condition is still relatively uncommon and a correct diagnosis is almost impossible. Some authors draw attention to the presence of a mass in the right iliac fossa in the early stages, appearing soon after the onset of symptoms. The case reported was complicated by diabetic coma, which added to the difficulties and probably delayed adequate treatment.

The two types of the disease, circumscribed and diffuse, are described and the respective treatment is discussed. In the case reported, exteriorization of the affected part of the caecum, utilizing the gangrenous part of the caecal wall for a caecostomy, was carried out successfully.

I wish to thank Mr. R. S. Ninian, senior surgeon, Maelor General Hospital, Wrexham, for permission to treat and publish this case, and also Dr. J. Forbes, physician, under whose care the patient was originally admitted.

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Medical Memoranda

Dextrocardia, Situs Inversus, and Twinning

A possible relationship between dextrocardia, situs inversus, and twinning has received some attention from geneticists. Thus, Ruud and Spemann (1922) produced situs inversus by division of the amphibian egg, the twin derived from the right half frequently showing this abnormality. This mirror-image mechanism, while a most attractive hypothesis, obviously does not occur in humans, as otherwise the condition would be relatively common. Reports of the incidence of situs inversus vary considerably, LeWald (1925) giving an incidence as low as 1 in 1,400, while Tanner-Cain and Crump (1951) quote an incidence of 1 in 35,000 in the United States Army recruits in 1947. Cockayne (1938) gives a figure of 1 in 19,500, which is probably more accurate, and is in agreement with most figures reported in the literature.

The purpose of this paper is to report an apparent association between dextrocardia, situs inversus, and a family history of twinning in ascendants and collaterals. This occurred in four out of five cases seen in general practice and school medical inspection. The first patient volunteered such a history, and investigation of three other cases, seen over a period of years, tends to confirm this finding.

Case Histories

Case 1.—A man aged 44. On routine examination in general practice in Lancashire a previously unknown situs inversus was diagnosed. This finding was subsequently confirmed by chest *x*-ray examination, screening, and barium meal. The mother and father were each one of twins.

Case 2.—A woman aged 48. Situs inversus was found on routine examination in general practice in Lancashire. The existence of this condition had been diagnosed previously as a result of an abdominal operation. Confirmation was obtained on *x*-ray and barium-meal examination. (*a*) The grandmother and grandfather were each one of twins. (*b*) A sister had twins. (*c*) An uncle had three sets of twins.

Case 3.—A girl aged 7. Previously unknown situs inversus was diagnosed at a routine school medical inspection in Lancashire. There was, in addition, a systolic murmur in the mitral area, suggestive of a small septal defect. This finding was confirmed by *x*-ray examination, screening, and electrocardiogram. It is of interest to record that her brother, aged 11, had a precisely similar systolic murmur in the mitral area, but no transposition of the viscera. (*a*) The father is a twin. (*b*) The paternal grandmother, twice married, had a set of twins by each husband.

Case 4.—A boy aged 6. Dextrocardia without reversal of viscera was diagnosed at a school medical inspection in Roscommon. There was, in addition, cyanosis of the lips and finger-tips, and a moderate degree of clubbing of the fingers and toes. A loud systolic murmur was audible at the base maximal to the right of the sternum, and a systolic thrill was also palpable. Pulmonary stenosis was the probable diagnosis. However, in view of the complicated necropsy findings in previously reported cases of isolated dextrocardia, it is possible that a more complex lesion may have been present. The diagnosis of dextrocardia was confirmed by *x*-ray examination and E.C.G. (*a*) The mother is a twin, probably monozygotic in type. (*b*) A full cousin, resident in America, has two sets of twins.

Case 5.—A girl aged 10. Previously unknown situs inversus was diagnosed at a routine school medical inspection in Lancashire. It was subsequently confirmed by *x*-ray examination and E.C.G. This was an adopted child, and it was not possible to investigate the family history.

Discussion

The exact genetic mechanism of situs inversus is in doubt. An association with twinning is mentioned by Doolittle (1907), who observed dextrocardia in a man who had a twin sister; both of his parents were one of twins, and he produced opposite-sexed twins, the boy showing dextrocardia. Foerster (1861) observed that in the majority of conjoined twins, particularly thoracopagi and pygopagi, one had complete or partial transposition of the viscera, and he advanced the theory that in cases of transposition there has been monozygotic twinning, with the subsequent death and absorption of the normal twin. This finding was not confirmed in the conjoined twins of Kano (Aird, 1954). In this instance there was dextrorotation of the heart in the twin surviving separation, but the E.C.G. was not that of a case of situs inversus. It was concluded, therefore, that there was no true mirror-image of the twins. Aird, however, states that 73% of conjoined twins show the latter abnormality.

In an extensive review of the subject, Cockayne (1938) stated that situs inversus is transmitted as a recessive characteristic caused by a single autosomal gene. He cited as proof for this theory, "its familial incidence and general distribution within a family, its occurrence in both members of a pair of monozygotic twins, and the high percentage of first-cousin marriages that give rise to it. In addition, the ratio of affected to normal sibs in the fraternities agreed with that expected of a recessive character." Twinning has long been admitted by both the medical and the actuarial professions to be a hereditary character, and if this association with twinning is accepted it tends to confirm that theory. However, there is some difficulty in explaining the occurrence of situs inversus discordantly in monozygotic twins. This was first reported by Dubreuil-Chambardel (1927) and has since been reported on a number of occasions, the most recent being that of Helweg-Larson (1947) in a pair of male twins. The normal twin produced one child and a pair of twins that miscarried.

Torgersen (1950) published details of 270 cases of situs inversus, discovered by mass radiography in Norway. He collected data concerning the familial occurrence of twinning in 100 unselected cases. He concluded that it did not

occur in any greater frequency than that expected in a normal population group. In a further investigation of families where the person with situs inversus was a twin, he concluded that the familial incidence of twinning was probably increased in this rare type of case. These findings are in conflict with my admittedly small series. However, the history of twinning in ascendants and collaterals in the families I have investigated is so striking that I suggest that this fact may be of importance genetically in some cases of situs inversus and is not mere coincidence.

I am indebted to Dr. M. L. Thomson, Royal Manchester Children's Hospital, and Dr. S. P. O'Toole, St. Patrick's Chest Hospital, Castlerea, for clinical investigation of Cases 3 and 4.

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A Case of Congenital Extra-abdominal Herniation of Bowel

The usual form of congenital herniation of abdominal viscera is into the base of the umbilical cord and is known as an omphalocele. An omphalocele has a large peritoneal sac which is not covered by skin. In the following case report the criteria necessary for the diagnosis of omphalocele are not present in that (1) the umbilical cord was normal and in no way connected with the hernia, and (2) the bowel had no parietal peritoneal covering. This case is recorded because standard works on the subject bear no description of such a congenital anomaly.

CASE REPORT

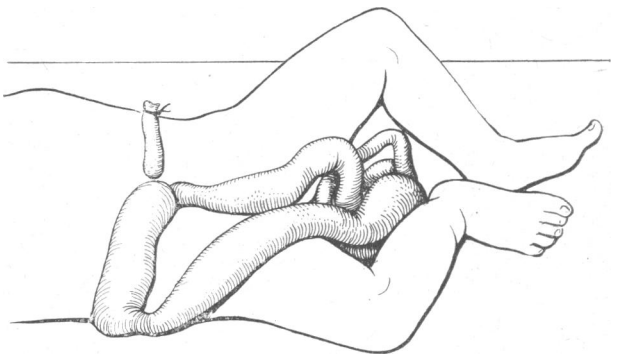
An infant was admitted to hospital on December 15, 1954, at the age of ½ hour. She was the first-born infant of parents both aged 28. The pregnancy had been uneventful; following spontaneous rupture of the membranes at thirty-four weeks a normal delivery resulted. Delivery was in a maternity home, and at birth the family practitioner (Dr. Philip Hogg) noticed the herniation of bowel and brought the infant immediately into hospital.

Examination revealed a newborn female baby in good general condition, although there was slight cyanosis of the lips. She cried lustily and was quite active. Her birth weight was estimated at 5 lb. (2.3 kg.). Physical examination revealed nothing abnormal apart from a cleft soft palate and the defect being described. The umbilical cord was normal, but 1 in. (2.5 cm.) to the right of the umbilicus, and separated by normal skin, was a clean-cut defect of the abdominal wall looking like a stab wound, through which approximately 2 ft. (60 cm.) of small bowel protruded (see illustration). There was no parietal peritoneal covering over the bowel and no hernial sac was present. The visceral peritoneum was thickened and the gut was brownish black in colour. Clinically there was doubt concerning the viability of the affected gut.

Operation.—At operation, by Mr. Stewart Feggetter two hours later, the 2 ft. (60 cm.) of thickened small gut was cleaned with saline and returned to the abdomen after the abdominal wall defect had been enlarged. The umbilical stump was removed and the abdomen closed in two layers.

Because of the excellence of the baby's general condition intravenous therapy was not given pre-operatively, and for the next few days she gave no cause for alarm. Intramuscular penicillin, 50,000 units six-hourly, and streptomycin, 50 mg. twice daily, were started immediately after operation because of the possibility of peritonitis developing. Gastric suction during the next three days produced minimal quantities of bile-stained fluid, and as she had had three small bowel actions by December 18 oral feeding was then begun for the first time. Hourly feeds of 1 dr. (3.5 ml.) of glucose-saline were given initially, and when this was taken reasonably well the amount was increased and she was changed over to half-cream national dried milk.

On December 24 vomiting started and her abdomen was seen to be distended. She had passed only one small motion in five days, and as a radiograph of the abdomen revealed the presence of fluid levels it was presumed that there was an obstruction. She was treated medically with intravenous fluids and gastric suction, and by December 26 the distension had disappeared and once more oral feeding was begun. From December 27 to 30 bowel washouts were



Drawing of the case, showing protrusion of small bowel.

given daily and normal results obtained, but on December 29 once again abdominal distension was noticed, but no vomiting occurred. The distension disappeared after her daily washout.

On January 1, 1955, she had her first spontaneous motion, and, as from then onwards she had spontaneous normal motions each day, the bowel washouts were discontinued. Her weight, which was 4 lb. 11 oz. (2.1 kg.) on December 19 and remained more or less stationary until January 1, started to increase, and on January 12 she was discharged weighing 5 lb. 11 oz. (2.6 kg.) and feeding satisfactorily.

On January 21 her mother reported that the infant had given "no trouble." Her weight had increased to 6 lb. 1½ oz. (2.8 kg.) in the nine days, and examination showed her to be a lusty normal baby. On February 22 her weight had risen to 8 lb. 15½ oz. (4.07 kg.). She was in excellent general condition, but a small ventral hernia was present at the lower end of her abdominal wound. When last seen, on March 22 at the age of 14 weeks, she weighed 11 lb. (5 kg.).

COMMENT

This form of hernia must be extremely rare, and, in view of the stab-like wound through which the gut protruded, one might consider the hernia to be traumatic in origin. However, the appearance of the gut suggested that it had been outside the abdominal cavity for a long time during intrauterine life. This suggested that its origin was congenital. Congenital abnormalities are often multiple, and the presence of a cleft soft palate seems to be further evidence in favour of its congenital origin.

With such a lesion, peritonitis and/or intestinal obstruction from adhesions is always a possible complication. Apart from the episode described above, this has not presented a problem to date, although it is possible that intestinal obstruction may develop in the future.

I thank Dr. Philip Hogg for the speed with which he brought the infant into our care, and Dr. George Davison and Mr. Stewart Feggetter for allowing me to publish this case history. I am extremely grateful to Dr. Davison for his helpful criticism.

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